

Portasystemic Shunting for Metabolic Disease

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For many years portal venous diversion has been used for the haemodynamic objectives of stopping or preventing haemorrhage from oesophageal varices or, less commonly, to treat intractable ascites. Since 1963, a new dimension has been added to the old operation of portacaval shunt by employing this procedure to alter favourably the course of patients with two inborn errors of metabolism, glycogen storage disease and hyperlipoproteinaemia. In this chapter we will discuss the results and the potential postoperative risks of portal diversion for these new indications, as well as the possible mechanisms of benefit.

GLYCOGEN STORAGE DISEASE

Portal diversion was first performed for glycogen storage disease almost 14 years ago.³⁴ Then the rationale for the procedure was naive by today's standards. It was hoped that by short-circuiting splanchnic venous blood around the liver, glucose would be made more readily available to peripheral tissues with relief of hypoglycaemia, and that the liver would be coincidentally deglycogenated. As will be discussed later, it has since been learned from animal experimentation that the consequences of portacaval shunt are far more subtle and wide-ranging than that simple view.

That first patient, who had Type III glycogen storage disease, is still alive almost 14 years after portacaval transposition. The by-passed portal venous blood is replaced with blood returning from the inferior vena cava with this operation, which was first described in animals by Child et al⁶ (Figure 45.1). The transposition was used in order to avoid the potential hazards of Eck fistula. It was appreciated then and amply confirmed since²⁴ that most animals, including subhuman primates, develop wasting and encephalopathy after portal diversion, but it was not appreciated that humans would be an exception to this generalization.

Our first patient had a remarkably untroubled convalescence and was discharged from the hospital nine days postoperatively. Before and after operation, there were elevations in the transaminases which a decade later were still slightly increased. She had a splenectomy 43 months after transposition for thrombocytopenia at which time the portacaval anastomosis draining the splanchnic bed was proved to be open. There has been no evidence of portal hypertension, indicating that the portacaval anastomosis draining the splanchnic bed is still open. An inferior vena cavagram almost 10 years postoperatively revealed flow of systemic venous blood from the distal vena cava to the liver but with a major by-pass around the liver through azygous and hemiazygous collaterals (Figure 45.2). The degree of azygous by-passing was similar to that observed with a

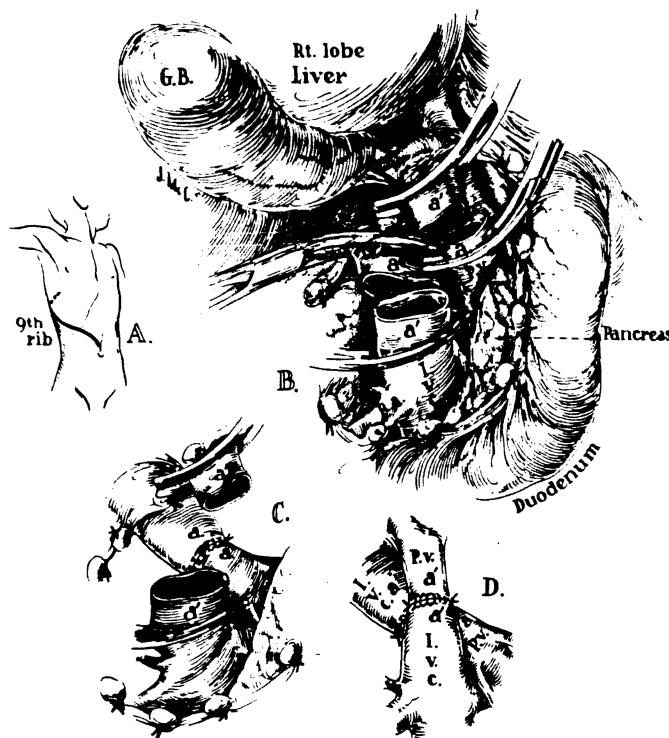


Figure 45.1. The operation of portacaval transposition that was used for the first two patients with glycogen storage disease who were treated with portal diversion. Note that the central portal vein is revascularized with vena caval blood. From Starzl et al.³⁴ with the kind permission of the editor of *Surgery*.

comparable angiographic study more than nine years earlier when the patient was three-months postoperative.

A patient similarly treated by Riddell et al²⁵ in Bristol, England, also survived for a long time. Riddell's patient moved to Canada and was temporarily lost to follow-up. Eventually he was traced through the Immigration Department. Angiographic studies were carried out in Calgary, Alberta, in December 1971, and confirmed by us in January 1973. The vena cava was obstructed (Figure 45.3, left), a complication which had probably occurred a long time previously since collateral venous channels were highly developed and lower limb oedema was minimal. The anastomosis draining the splanchnic venous bed was shown to be open (Figure 45.3, right).

One more portacaval transposition was performed. The attempt cost the life of our second patient, when the liver was unable to transmit the rerouted vena caval flow, causing hepatic swelling and uncontrollable acidosis. This seven-year-old child died two days later. We²⁷ and Hermann and Mercer¹⁶ subsequently recommended that the simpler procedure of portacaval shunt be used. To our knowledge this approach has been followed in all later

cases. By the spring of 1973³⁵ our own series of patients had reached seven, and six more had been formally reported in the literature from other centres. Since then our cases have increased to 10 and it is upon this personal experience (Table 45.1) that most of the opinions in this report will be based. We have had no operative deaths or technical failures after end-to-side portacaval shunt.

Our original patient with transposition is still living after 14 years, as are seven of the eight patients treated with simple portacaval shunts. These last seven patients now have follow-ups of one to 5½ years (mean 50 months). The ages of all of our 10 patients, types of disease and symptoms are summarized in Table 45.1. Type I disease (glucose-6-phosphatase deficiency) has been the most common indication for treatment, with Type III disease (amylo-1,6-glucosidase deficiency) being a distant second.

Metabolic effects

After portal diversion, most of the children who had pre-existing hypoglycaemia did not have relief of this problem or the relief was not complete. Thus, night feedings usually had to be

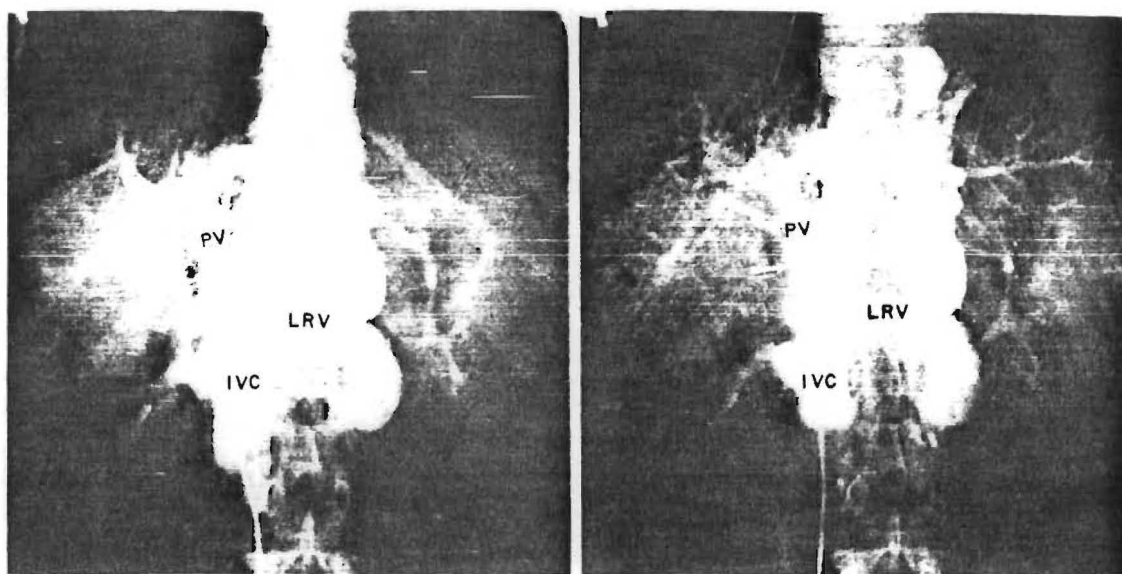


Figure 45.2. Inferior vena cavagram in March 1973, 9½ years postoperatively in Colorado Case 1 showing a patent caval-portal anastomosis (arrow) under two conditions of dye injection. Significant flow through the liver, as well as around it by the azygous and other collaterals, was well seen at fluoroscopy. IVC - distal inferior vena cava; LRV - left renal vein; PV - portal vein. From Starzl et al.³⁵ with the kind permission of the editor of *Annals of Surgery*.

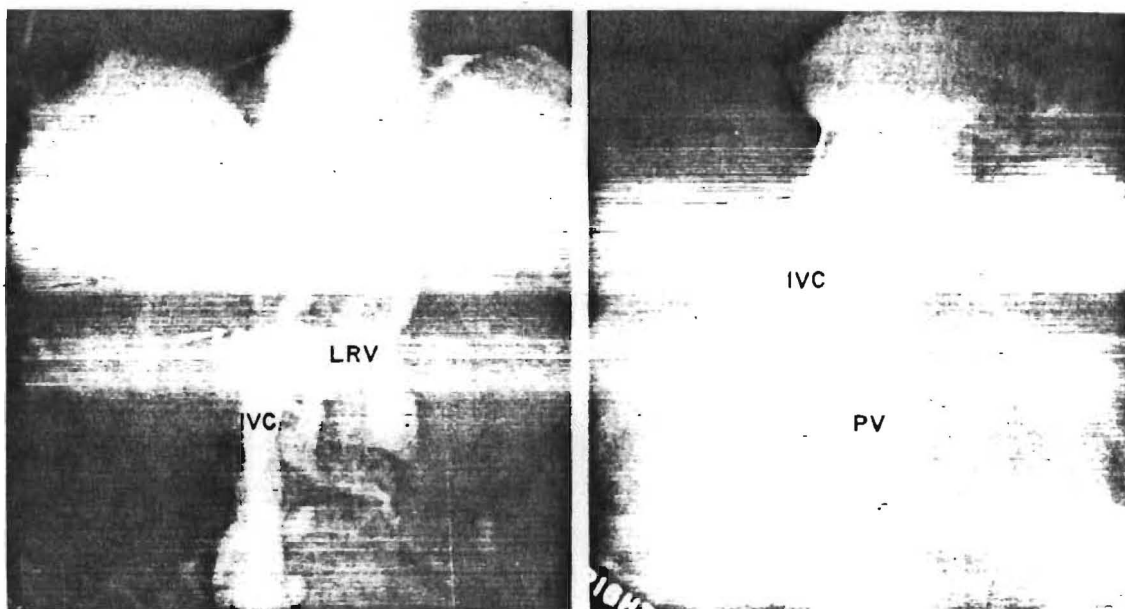


Figure 45.3. Studies of the Bristol case of portacaval transposition. The original operation was on 5 May 1965, and the examination depicted, which was in December 1971, was performed by Dr R.J. Sommerville at the Foothills Hospital, Calgary, Alberta. (Left) Inferior vena cavagram showing obstruction at the anastomotic site (arrow). IVC - distal inferior vena cava; LRV - left renal vein. Note the extensive collaterals via the azygous system. (Right) Demonstration of a patent portacaval anastomosis (arrow) by means of a retrograde catheterization. IVC - proximal inferior vena cava; PV - portal vein. From Starzl et al.³⁵ with the kind permission of the editor of *Annals of Surgery*.

Table 45.1. Patients with Glycogen Storage Disease Treated by Portal Diversion at the University of Colorado.

Case No.	Age	Glycogen storage disease type	Date of operation	Symptoms preoperatively			Persistent hypoglycaemia postoperatively	Survival post-shunt
				Hypo-glycaemia	Acidosis	Growth retardation		
1 ^a	8	III	10.15.63	x	x	x	No	Alive 14 years
2 ^a	7	I	6.26.68	x	x	x	-	Died 2 days
3	7	I	5.2.72	x	x	x	Yes ^b	Alive 5 years
4	11	I	5.17.72	x	x	x	No	Died 4½ years
5	10	VI	8.2.72			x	-	Alive 4½ years
6	5	III	11.7.72	x	x	x	No	Alive 4½ years
7	3	III	11.8.72	x	x	x	Yes	Alive 4½ years
8	8	I	8.13.73	x	x	x	Yes ^b	Alive 3½ years
9	12	I	12.14.73	x	x	x	Yes ^b	Alive 3½ years
10	1	I	9.2.76	x	x	x	Yes	Alive 8 months

^a Operation was portacaval transposition. The last eight patients had simple portacaval shunt.
^b Overnight feeding via nasogastric tube started 2½ to 4 years after portacaval shunt.

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r, as well as around it
left renal vein: PV



5 May 1965, and the
Foothills Hospital.
IVC - distal inferior
cav: PV - portacaval

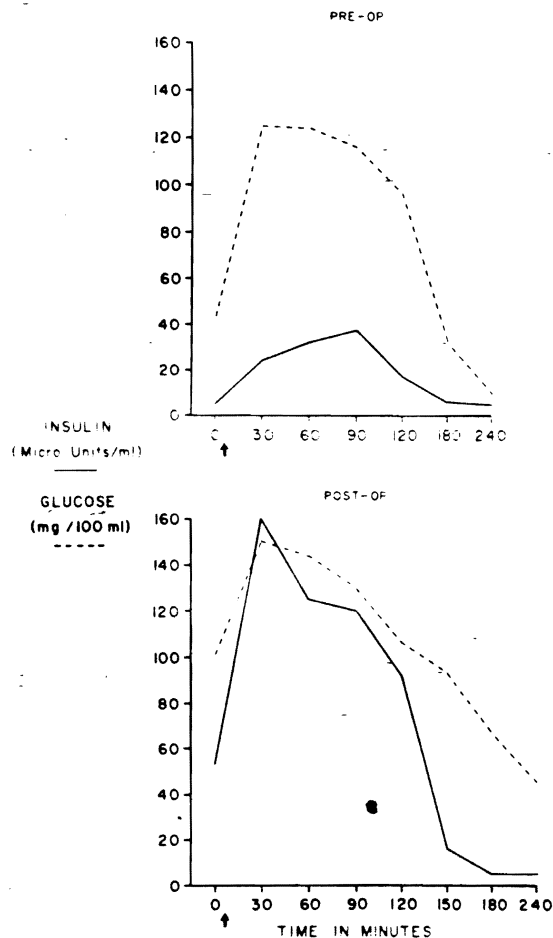


Figure 45.4. Plasma insulin and glucose concentrations before and after portacaval shunt in a child (Case 8, Table 45.1) with Type I glycogen storage disease. Oral glucose tolerance test.

continued. Studies of plasma insulin and glucagon in several of these patients have revealed changes (Figure 45.4). The flat peripheral insulin curves typical of Type I glycogen disease²² became significantly elevated after portacaval shunt, and there were smaller increases in glucagon. The glucose tolerance curves were a little different before and after operation.

Liver glycogen concentrations in all the patients later biopsied were not changed. In spite of this, the liver size in several of our patients and those reported by others underwent a very obvious reduction as measured with liver scan planimetry. Even if obvious shrinkage did not occur, postoperative biopsies always showed a diminution in individual hepatocyte size similar to that produced in animals by portacaval shunt.³⁵

In contrast to the incomplete relief of hypoglycaemia, all components of the hyperlipidaemia which is a characteristic of the Type I disease had profound and permanent relief (Figure 45.5), as was first observed by Hermann and Mercer¹⁶ and confirmed by Folkman et al¹² and in our own cases.³⁵ Correction of other metabolic defects was observed, including abnormal bleeding, uric acid elevations and abnormal calcium metabolism.³⁵

Growth

All 10 of our patients had growth retardation before portacaval shunt. Afterwards, height

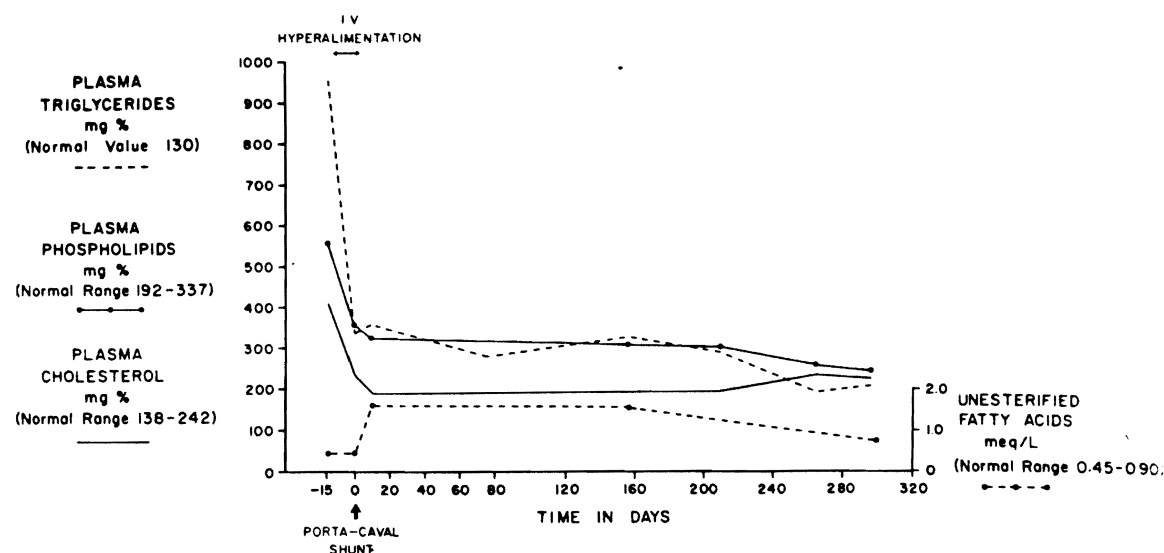


Figure 45.5. Effect of parenteral hyperalimentation and end-to-side portacaval shunt on the plasma lipids of Patient 4 whose diagnosis was Type I glycogen storage disease. Note the rapid and relatively complete reversal of all abnormalities. From Starzl et al,³⁵ with the kind permission of the editor of *Annals of Surgery*.

increases, which in most cases had virtually ceased, have occurred at the rates listed in Table 45.2, that is, approximately 0.5 cm per month.

Quantitative measures of growth were obtained with radiographic techniques.³⁵ An example of the results is shown in Figure 45.6. Comparison of the wrist and hands in this seven-year-old stunted child before and 11½ months after operation showed the phenomenal effects of bone age doubling. In addition to the size change, mineralization occurred, as did the

appearance of new wrist bones. Circulating somatotrophin in these patients was normal. The growth spurts may have been at least partially attributable to the increased insulin distribution to the periphery mentioned earlier (see Figure 45.4) since, in recent years, insulin has been recognized to be a major growth hormone comparable in potency to somatotrophin. The simpler possibility that better nutrition was responsible must also be considered.

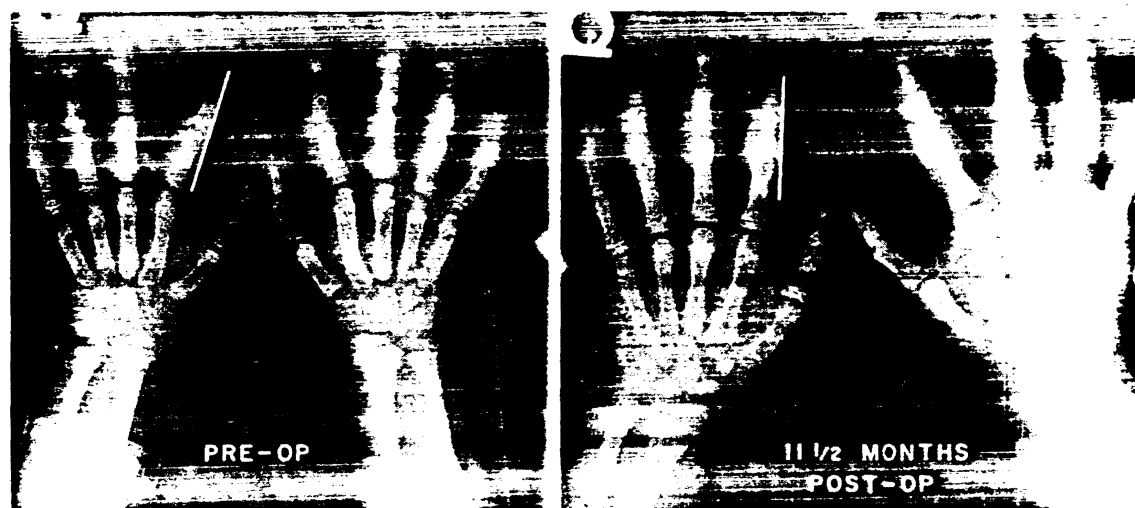


Figure 45.6. The dramatic wrist and hand bone growth and mineralization in Case 3 in the first 11½ postoperative months. The bracket on the left index finger is 5 cm in length. From Starzl et al,³⁵ with the kind permission of the editor of *Annals of Surgery*.

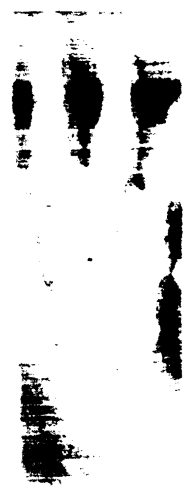
Table 45.2. Growth Rate and Complications Following Portal Diversion for Glycogen Storage Disease.

Case No.	Growth rate (cm/month over 9 to 120 months)	Complications
1	0.49	
2	Op. Death	
3	0.50	Macroadenomatosis
4	0.28	Died 4½ years post-shunt. Primary pulmonary hypertension. NH ₃ = 85 µg/100 ml; macroadenomatosis
5	0.53	Renal artery stenosis surgically corrected 32 months post-shunt
6	0.62	
7	0.50	
8	0.49	
9	0.88	Renal stone two months post-shunt. Mild arterial hypertension; macroadenomatosis
10	0.22	
Mean	0.47	

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Encephalopathy and other risks

None of our patients has developed hepatic encephalopathy. The highest blood ammonia recorded was 49.3 µmol/l (84 µg/per cent) in a child who died almost five years after portacaval shunt during an attempt at transcaval radiographic visualization of the portacaval anastomosis. This patient (Case 4) had not achieved the full expected growth after operation (Table 45.2), had mild systemic hypertension, and had unexplained transient bouts of cyanosis, hyperventilation and unconsciousness. Except for the slightly elevated blood ammonia, liver function was normal.

At autopsy, the liver had macroadenomatosis. Very minor protoplasmic astrogliosis was present in the brain. A finding that had not been suspected during life was advanced right ventricular hypertrophy and dilatation. The smaller pulmonary arteries and arterioles had medial muscle hypertrophy, medial and intimal fibrosis, scattered fibrinoid necrosis and numerous plexiform lesions. In retrospect, the patient died from a cardiopulmonary complication of the kind that has been documented in patients with Type I glycogen storage disease or other liver disease.²¹

This complication has been termed 'vasoconstrictive pulmonary hypertension'. It has been speculated that a humoral vasoconstrictive agent, which normally is completely detoxified

by the liver, is responsible for the hypertrophic lesions in the pulmonary vasculature. It is conceivable, although unlikely, that the development of a renal artery stenosis in Patient No. 5 (Table 45.2) could have been caused by the same mechanism.

Three of our 10 patients developed the hepatic lesions which have been termed both macroadenomatosis and nodular hyperplasia. Filling defects were noted by liver scan with some waxing and waning of size. This complication is common in patients with Type I glycogen storage disease, particularly with advancing age, and was recently reported in seven of eight patients who were three to 28 years old.¹⁸

The value of enteral or parenteral feeding

Seven years ago, Folkman et al¹² of Boston Children's Hospital added an important therapeutic dimension by showing how preoperative parenteral hyperalimentation would reduce the risk of portacaval shunt by normalizing pre-existing hepatomegaly, acidosis and other abnormalities, including hyperlipidaemia. The Harvard group as well as the Vanderbilt team^{3,14} have greatly extended these original observations.

Greene et al,¹⁴ for example, showed that continuous night feeding through a gastrostomy or gastric tube resulted in many of the same benefits as the parenteral hyperalimentation or as the portacaval shunt. Apparently, the night feeding was originally tried to control the nocturnal hypoglycaemia which, as mentioned earlier, was not relieved by portal diversion. It was soon observed that the night feedings contributed to, or were primarily responsible for, the same kinds of growth spurts and relief of metabolic abnormalities (including hyperlipidaemia) which are ameliorated by portacaval shunt. Greene et al¹⁴ have shown that systemic plasma insulin levels are more than doubled by such treatment and that glucagon is decreased, leading them to speculate that the hormone changes were responsible for the benefits. Whatever the explanation, this kind of treatment is an alternative to portal diversion as the primary treatment of children with glycogen storage disease, and even after portacaval shunt it has an additional value as has been evident in three of our patients (Table 45.1) who had hypoglycaemia, retardation of growth, or both. Glucose and amino acid mixtures have both been effective for the gastric feeding.

HYPERLIPIDAEMIA

The observations about lipid lowering after portacaval shunt in patients with Type I glycogen storage disease (see Figure 45.5) were responsible for our suggesting the same operation to treat hyperlipidaemia.³¹ It has been 4½ years since a completely diverting portacaval shunt was first performed clinically for the purpose of lowering the serum concentration of cholesterol and low density lipoproteins (LDL). Since then, we have treated two more patients by this method. As of May 1977, an additional nine patients had the same procedure in other centres and were documented in the literature.^{8, 11, 20, 26, 36, 37}

These 12 cases represent less than half of the true total. Through an informal registry that has been kept at the National Institutes of Health, Bethesda, U.S.A., it is known that more than 30 portacaval shunts have been carried out for the treatment of hyperlipidaemia with results that are in general agreement with the smaller sample which is accessible through the formal literature and upon which we will comment in this chapter. All three of our patients had homozygous Type II hyperlipidaemia.

In Type II hyperlipidaemia, Goldstein and Brown¹³ have suggested that there may be an absence of or defect in the cell surface receptor sites which normally bind and transport LDL cholesterol into the cell. Because cholesterol does not enter the cell, they proposed that there is an absence of the normal feedback suppression of cholesterol synthesis.

The homozygous form of Type II hyperlipoproteinaemia has a shockingly poor prognosis-if, as is usually the case, there is not a good response to medical therapy. Lipid-rich deposits are laid down in widely separated superficial and deep parts of the body, including blood vessels where premature atherosclerosis is the consequence. Cardiac valves are similarly affected, with aortic stenosis being particularly common. The patients usually die of cardiovascular complications before the age of 20 years despite all conventional therapy. The gravity of the situation seemed to justify the trial of a new approach.

Colorado cases

Our first patient, an 11-year-old girl, had suffered a major myocardial infarction about

two months before the portacaval shunt. After the portal diversion, her serum cholesterol values fell from about 20 mmol/l (800 mg/dl) to levels that were consistently below 10 mmol/l (400 mg/dl) (Figure 45.7). Unsightly xanthomas began to resorb from visible subcutaneous and tendinous locations (Figure 45.8). Attacks of pre-existing angina pectoris became less frequent and finally stopped. By cardiac catheterization 16 months after the end-to-side portacaval shunt was performed, there was good evidence that reversal of aortic stenosis had occurred, with a diminution of the aortic valve gradient from 7.45 to 1.33 kPa (56 to 10 mmHg).²⁸ The coronary arteries were also thought to be less diseased than before, although three stenoses were still present. More than 1½ years after portacaval shunt, the girl died suddenly while coming home from school.

At autopsy, the residual coronary artery disease was confirmed. There was a large ventricular aneurysm. The conclusion was that death was probably caused by an acute cardiac arrhythmia.³³ She should have been submitted to a coronary revascularization procedure as was done in Case 2 (see below). The decision against this was taken with the hope that the coronary artery stenoses would regress spontaneously.

The portacaval shunt was widely patent. The liver, which weighed 618 g, was grossly normal, and microscopically it was unchanged from the biopsy specimen that was obtained six months postoperatively. On light and electron microscopy, the most prominent findings were shrinkage of the hepatocyte size, depletion of rough endoplasmic reticulum, and the accumulation of intracytoplasmic lipid deposits. These changes are typical of those caused in the liver

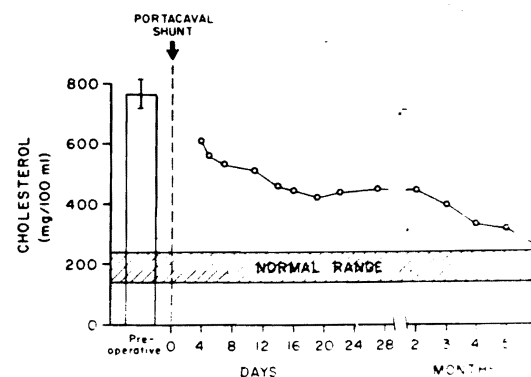


Figure 45.7. Serum cholesterol concentrations after portacaval shunt in Patient 1. Table 45.3.

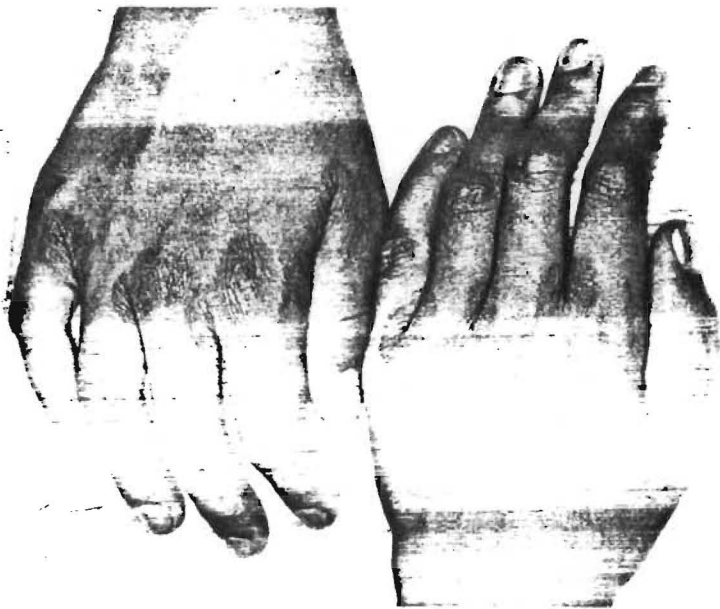
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(a)



(b)

Figure 45.8. The hands of Patient 1 of the hyperlipidaemia series two weeks before (a) and 16 months after (b) portacaval shunt.

by portacaval shunt in all species studied so far.²⁴ Hepatic function had not been changed by portacaval shunt as judged by standard liver function tests.

We have performed a portacaval shunt on two more patients with the same diagnosis (Cases 2 and 3, Table 45.3). Patient No. 2 was a seven-year-old girl at the time of operation. She had preoperative serum cholesterol values that averaged 25.82 ± 1.22 (s.d.) mmol/l (997 ± 47 [s.d.] mg/100 ml) while she was on a very low

cholesterol diet. Six months after the shunt, the cholesterol level measured in the same laboratory was 15.54 mmol/l (600 mg/100 ml) (a 40 per cent reduction), despite a relaxation of the diet. These values have continued to fall under careful medical management by Dr David Bilheimer of Southwestern University Medical Center. The cholesterol concentrations during the last year have been 11.66 ± 1.73 mmol/l (450 ± 67 mg per cent). The xanthomas on her heels, elbows and hands either have resorbed

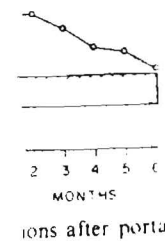


Table 45.3. Patients with Type II Hyperlipidaemia Treated at the University of Colorado.

Case No.	Age	Date of operation	Serum cholesterol mmol/l (mg/100 ml)			Clinical result	Comments
			Pre-shunt	Post-shunt	Final % reduction		
1	12	3.1.73	19.92(769)	7.51(290)	62	Died 18 months; cardiac arrest	Old myocardial infarct with ventricular aneurysm; residual stenoses of coronary arteries; xanthomas had disappeared
2	7	10.4.74	25.82(997)	11.66(450)	55	Good	Had aortic and mitral valve replacement plus double coronary artery by-pass 25 months after shunt; xanthomas nearly completely disappeared
3	8	8.5.75	25.90(1000)	15.54(600)	40	Good	Xanthomas slowly disappearing

completely or have greatly diminished in size.

This child also had aortic stenosis and angina pectoris. Because of persistence of her cardiac disease, she was treated with aortic and mitral valve replacement plus double coronary artery by-pass two years after portacaval shunt. The operation was carried out by Dr Robert McGehee and his associates at the Fort Worth Children's Hospital. The patient has returned to school with essentially unlimited activities.

Dr Bilheimer has written us: 'Our therapeutic approach continues to be one of trying to get Mary's cholesterol as low as possible with medication. I am totally convinced that the portacaval shunt was tremendously helpful to her. I doubt that we would have proceeded with the cardiac surgery if her cholesterol were still in the 23 to 25 mmol/l (900 to 1000 mg per cent) range. Her present cardiac difficulties are an extension of those already present before the shunt surgery. Our main job now is to try and retard the development of atherosclerosis in her coronary by-pass grafts.'

A third patient had a portacaval shunt performed 24 months ago. The child was from France and has been managed through a collaborative effort with Professor Jean Rey and Dr J. Schmitz of the Hôpital des Enfants Malades and Professor J.-L. deGennes of the Hôpitaux de Paris (Clinique Endocrinologique de la Pitié). The patient was eight years old at the time of operation on 5 August 1975. Preoperatively, typical xanthomas were prominent on the feet, knees, elbows, buttocks and popliteal fossa. The serum cholesterol concentration averaged 25.90 mmol/l (1000 mg per cent) with very little variation despite a low

cholesterol diet and maximal treatment with cholestyramine, nicotinic acid and clofibrate. Skin culture results¹³ were diagnostic of homozygous Type II hyperlipidaemia. Aortic angiography in April 1975 showed severe atheromatous disease of the aorta with supra-aortic narrowing. The coronary, mesenteric and renal arteries were patent. There was mild aortic insufficiency.

After portal diversion the serum cholesterol value fell to 14.3 to 16.8 mmol/l (550 to 650 mg per cent) (40 per cent fall). The visible xanthomas slowly resorbed. The patient was asymptomatic before operation and remained so afterwards with good growth and development. His working electrocardiogram was normal in June 1977. At that time, mesenteric arteriography was performed at the Hôpital de la Pitié in Paris. Late phase venography showed the portacaval shunt to be patent.

The patient was seen in Denver during the first week of August 1977. The cholesterol concentrations ranged from 15 to 16 mmol/l (580 to 616 mg per cent). Liver function tests were normal except for a blood ammonia of 93 μ mol/l (158 μ g per cent) (normal less than 35 μ mol/l [less than 60 μ g per cent]).

Changes in peripheral insulin and glucagon content in two of our three patients have been noted. Preoperative values were normal. Post-operatively, systemic venous insulin and glucagon values were both increased, especially the former (Figure 45.9).

None of our three patients with hyperlipidaemia has had any signs of hepatic encephalopathy, although one (Patient 3) has an elevated blood ammonia value. Nevertheless

Pt. 2

— Pre Shunt
 --- Post Shunt

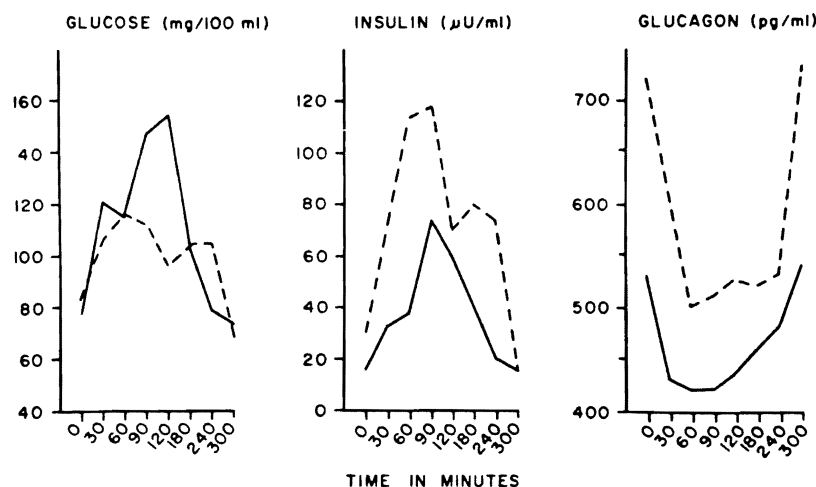


Figure 45.9. Systemic venous plasma glucose, insulin and glucagon concentrations in Patient 2 with hyperlipidaemia before and after portacaval shunt. Oral glucose tolerance test.

the use of portacaval shunt to ameliorate hyperlipidaemia accepts a trade-off of sub-optimal conditions of liver perfusion in return for metabolic improvements (see 'Mechanism of Portal Diversion Effects', below). Realization of this fact has prompted us to recommend portacaval shunt for this inborn error only if the patient was homozygous. However, the freedom from encephalopathy in our cases suggests that symptomatic heterozygotes who are refractory to medical therapy should also be considered for portal diversion. The patient of Weglicki et al.³⁷ probably had the heterozygous trait.

Other reports

In the nine cases reported in the literature by others,^{8,11,20,26,36,37} the decreases in serum cholesterol have been variable from patient to patient, as in our three cases. A significant fall was usually obtained if the shunt was patent. Cardiac symptoms were often relieved and resorption of xanthomas proceeded even if the falls in serum cholesterol concentration were not great. The latter finding has raised the possibility that cholesterol depots may keep the serum cholesterol concentrations high until the resorption is nearly complete. Failure to observe an antilipidaemic effect has been associated with thrombosis of the shunt.^{8,11}

Weglicki et al.³⁷ have used portacaval shunt and coronary artery by-pass together in a young patient suffering from premature atherosclerosis. This kind of combined therapy will probably be used more frequently in the future, as was illustrated also by the management of our Patient 2.

MECHANISM OF PORTAL DIVERSION EFFECTS

It has been increasingly appreciated during the last decade that venous blood from the splanchnic viscera has liver-supporting qualities not found to the same degree in other kinds of arterial or venous blood.^{29,30,32} The so-called hepatotrophic effects of portal blood have been noted under several experimental conditions to include hypertrophy, glycogen storage, hyperplasia and increase of several synthetic functions. The main splanchnic venous hepatotrophic factors are almost certainly endogenous hormones of which the single most important seems to be insulin. The high concentrations of nutrients from the intestine probably play a significant contributory role.

With portacaval shunt these humoral and non-humoral portal factors are lost to the liver on first pass. Subtle but fundamental changes in

hepatocyte structure and function result. As suggested earlier, changes in the hormone environment of the liver and the rest of the body caused by portacaval shunt could explain the palliation of patients with glycogen storage disease.

The loss of portal hepatotropic factors may also explain the amelioration of hyperlipidaemia. The antilipidaemic effect of portacaval shunt can be demonstrated with a normal starting level of serum cholesterol just as easily as with a pathologically high concentration. This was first observed by Winter et al³⁸ in normal dogs and confirmed in that species by us³² and by others.^{7,15,17} The same has been seen in rats,^{10,23} pigs^{4,5} and baboons.²⁹

Our studies in the dog²⁹ and investigations in the rat,^{19,23} pig^{4,5} and human² have shown or suggested that a reduction in hepatic cholesterol and/or LDL synthesis is responsible, at least in part, for the cholesterol falls. Only the report of Coyle et al in dogs has denied this.⁷ Other factors may contribute to the antilipidaemic effect of portal diversion as Ahrens and we have speculated.^{1,29}

Our experiments in dogs have suggested that the main reason for decreased hepatic cholesterol synthesis is deprivation in the liver of insulin.²⁹ One consequence is a moderately severe liver injury of the kind described earlier in the autopsy findings of Patient 2.

Eaton⁹ has recently proposed that the changes in glucagon metabolism may be important in the antilipidaemic effect of portal diversion. Further clarification of these mechanisms will be important if treatment alternatives to portacaval shunt are to be evolved.

SUMMARY

Complete portacaval shunt was used to treat 10 patients with glycogen storage disease. A favourable effect was noted on body growth and a number of metabolic abnormalities. More recently, continuous night feedings with an intermittently-placed gastric tube or through a gastrostomy has been shown to be helpful either before or after portacaval shunts. Such alimentation techniques may eliminate the need for shunts in some patients and be of adjuvant benefit in others.

Three patients with homozygous Type II hyperlipidaemia were submitted to complete

portacaval shunt. There was a substantial fall in serum cholesterol in each case, resorption of xanthomatous deposits on the skin and tendons, and possibly a partial reversal of some cardiovascular complications. One patient died, 18 months after portacaval shunt, apparently from a cardiac arrhythmia. The other two patients are in good health, one after subsequent double heart valve replacement and double coronary artery by-pass. Portal diversion seems capable of markedly altering the outlook of patients with homozygous Type II hyperlipidaemia.

Experience with portal diversion to treat patients with glycogen storage disease and hyperlipidaemia has also been accumulated in other centres with generally confirmatory results. If the desired corrections are not realized, thrombosis of the portacaval shunt must be suspected.

ACKNOWLEDGEMENTS

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